HEREDITARY MOTOR AND SENSORY NEUROPATHY MIMICKING HANSEN'S DISEASE

V K Somani, V Sucharita, V K Sharma, V N V L Sita, Fatima Razvi

A rare case of hereditary motor and sensory neuropathy in a 45-year-old man with glove and stocking hypoaesthesia, bilateral clawing, foot drop, and thickening of the peripheral nerves mimicking Hansen's disease is reported.

Key Words: Hereditary motor and sensory neuropathy, Hansen's disease

Introduction

Hereditary motor and sensory neuropathy (HMSN) is a rare syndrome, often familial, characterised by enlargement of peripheral nerves, associated with symptoms of slowly progressive polyneuropathy, and often with other abnormalities. Peripheral nerves show recurrent demyelination and remyelination with Schwann cell proliferation. Three separate types (I,II and III) are described.[1]

Case Report

A 45-year-old male patient presented with difficulty in walking and high stepping gait of 33 years duration, weakness and deformity of upper limbs of 16 years duration and a trophic ulcer on the right great toe of 1 week duration. His grandfather had suffered from a similar condition.

There was no evidence of hypopigmented patches anywhere on the body. There was hypoaesthesia in a glove and stocking distribution over the acral parts of the limbs. There was thickening of left ulnar and left lateral popliteal nerves, and gross wasting of the interossei, hypothenar and thenar muscles on both sides with bilateral clawing of the hands (Fig.1) and bilateral foot drop. There was a trophic ulcer on the right great toe (Fig.2). Ankle jerks and plantar reflexes were absent on both sides and the other reflexes were sluggish.

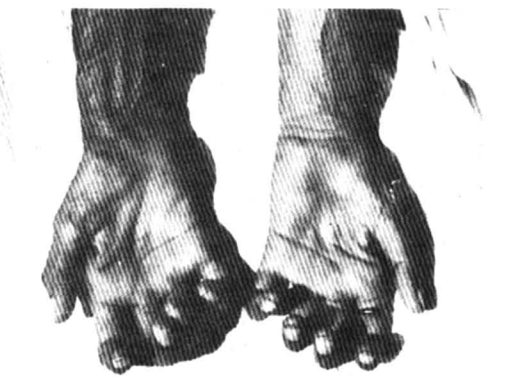

Fig. 1. Bilateral clawing, wasting of the thenar, hypothenar and forearm muscles.

Routine investigations like haemogram, blood sugar and urea were within normal limits. Skin smears from six sites and also nasal blow did not show acid fast bacilli. Lumbar puncture was normal. Nerve conduction studies revealed prolonged distal latencies, reduced compound motor action potential (CMAP) amplitudes and conduction velocities of the right median and right ulnar nerves. The F-responses were absent. In the lower limbs the CMAPs of the peroneal and tibial nerves bilaterally could not be elicited. The sensory nerve action potential (SNAP) amplitudes of the right median, ulnar and

From the Department of Skin and VD, Deccan College of Medical Sciences, Princess Esra Hosptial, Shah Ali Bunda, Hyderabad, India.

Address correspondence to : Dr V K Somani 2-1-43/A, Nalla Kunta, Hyderabad - 500044.

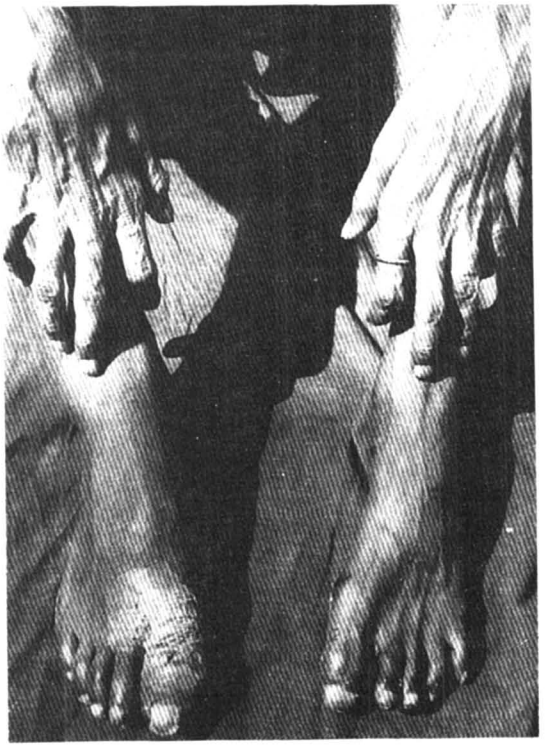

Fig. 2. Bilateral clawing and foot drop with wasting of the interossei of hands and feet and a trophic ulcer on right great toe.

both sural nerves could not be elicited suggestive of demyelinating neuropathy and features of significant reinnervation suggestive of long standing disease.[2]

Discussion

There are only a few diseases which can match the spectrum of clinical features exhibited by Hansen's disease. Referral to our department was for exclusion of leprosy as the telltale signs of bilateral acral hypoaesthesia, mimicking the glove and stocking distribution of leprosy, muscular atrophy, clawing of the hands, bilateral foot drop and thickening of the nerves with a trophic ulcer were all present to justify the clinical impression of leprosy.

However, closer questioning and examination revealed a family history, typical age of onset, slow characteristic progression of the disease, including shooting pains, difficulty in walking and a high stepping gait with foot drop[3] and enlarged nerves aroused a clinical suspicion of HMSN. The signs of central nervous system involvement ie, loss of reflexes further excluded the possibility of leprosy.[4] Nerve conduction studies revealed that HMSN was of type I. Skin biopsy was unremarkable.

So, though rare, HMSN is to be considered when a case clinically manifesting as lepromatous leprosy but smear negative presents, because it very closely mimicks the features of leprosy.

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